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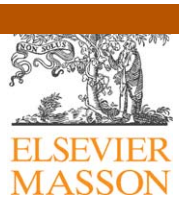

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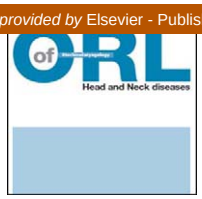

CLINICAL COMMENTARY

A case of middle-ear angiosarcoma

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KEYWORDS
Angiosarcoma;
Middle-ear;
Large tumor removal

Summary
Introduction: Middle-ear angiosarcoma is a rare malignant tumor that requires early diagnosis to allow satisfactory removal. The objective of the present study was to describe and discuss the diagnosis and treatment of this rare malignant middle-ear tumor.
Case report: A 12-year-old girl was admitted for a left retroauricular mass and a budding left external auditory canal tumor without associated peripheral facial nerve palsy. CT scan showed a mastoid-region tumor extending to the external auditory canal with mastoid and occipital bone-loss. Tumor removal used a retroauricular approach. Postoperative diagnosis was of angiosarcoma. At 12 months' regular follow-up, there were no signs of local recurrence.
Discussion and conclusion: Middle-ear angiosarcoma is a rare tumor, in which diagnosis is late because early symptoms are not specific. A multidisciplinary approach is essential for efficient management.

Introduction

Malignant tumor of the ear is infrequent, with an incidence of 1 per six million of the population. Whatever the histological type, a middle-ear location is exceptional [1]. Middle-ear angiosarcoma is thus rare. Diagnosis should not be delayed, so as to allow satisfactory tumor removal without risk of neurologic sequelae. Prognosis is generally poor but depends on the time to diagnosis [2]. The present observation provides an opportunity to describe and discuss the diagnostic and therapeutic management of middle-ear angiosarcoma.

Case report

A 12-year-old girl was admitted for chronic unilateral left otorrhea with 2 years' evolution, associated with retroauricular tumefaction. Clinically, she presented with a budding tumor filling the external auditory canal and a firm retroauricular mass of 6 cm diameter (Fig. 1). There was no associated peripheral facial nerve palsy. CT scan confirmed the presence of a mastoid-region tumor extending to the tympanum and external auditory canal. There was osteolysis of the mastoid and posterior external auditory canal wall (Fig. 2). Removal via a retroauricular approach used conchoplasty (Fig. 3). Immediate postoperative course showed surgical site suppuration despite systematic empiric antibiotherapy. There was also grade-3 facial nerve palsy (House and Brackmann classification). The surgery site infection

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Figure 1 Retroauricular mass and external auditory canal.

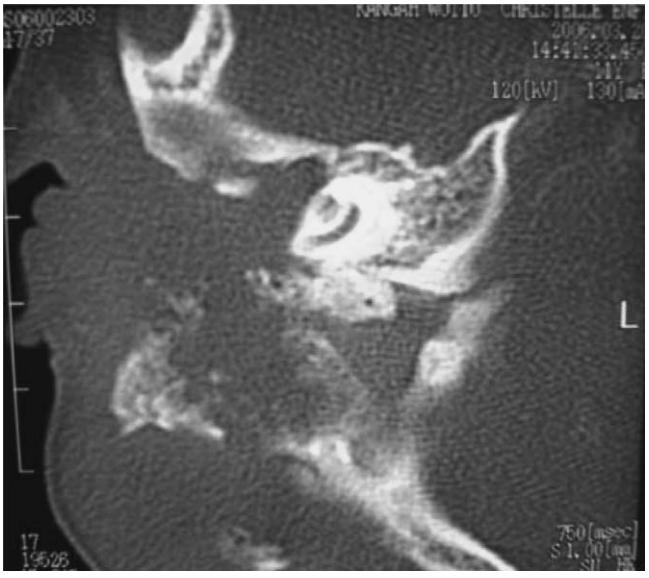

Figure 2 Axial CT slice showing mastoid osteolysis.

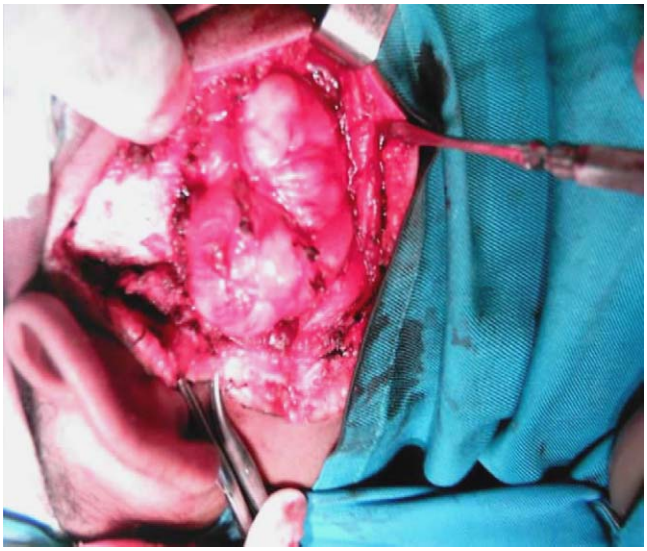

Figure 3 Mastoid-region tumor extending to external auditory canal by osteolysis of the posterior canal wall.

was arrested by adapted antibiotherapy, and the facial palsy recovered fully. Histologic examination of the surgical specimen found several vascular fissures with endothelial cell nucleus abnormalities, establishing the diagnosis of middle-ear angiosarcoma extending to the external auditory canal (Fig. 4). Complementary radiochemotherapy was prescribed but could not be performed, radiotherapy not being available in Côte d'Ivoire. The patient was referred to the oncology department, where chemotherapy was administered. At 12 months' follow-up, there was no sign of recurrence.

Discussion

Primary malignant tumors of the middle-ear are rare, representing 5% of ear cancers [1]. Eighty percent of middle-ear tumors are in fact extensions of outer-ear neoplasia, notably of the external auditory canal [2]. In the present case, the primary site was the mastoid, with secondary extension to the tympanum and external auditory canal by lysis of the posterior canal wall. Mean age at onset depends on the

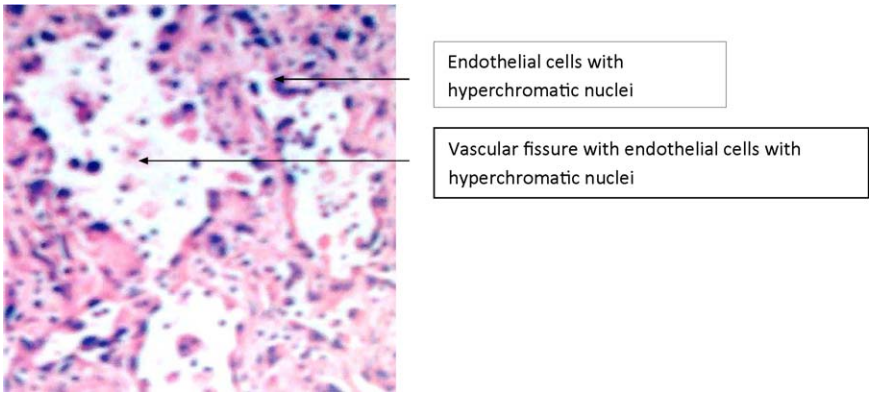

Figure 4 Histologic cross-section (HEX 100) showing several vascular fissures with cytonuclear abnormality of the endothelial cells.

Table 1 Middle-ear angiosarcoma cases in the literature.

Authors	Country	Number
Cernyl [7]	Czechoslovakia	1
Fishbun [8]	Russia	1
Alvares et al. [1]	Brazil	1
Present case	Côte d'Ivoire	1

histologic type [2,3]: for sarcoma, onset is during the first decade of life [4]. Chronic otitis is a sarcoma risk factor [1,2,3,5]. The present patient presented with chronic otorrhea, which in our opinion was rather a consequence of the tumor. It is, however, nonspecific to malignant ear tumor, inducing error in diagnosis, which accounts for the long delay (24 months) in treatment in the present case. Moffat et al. [5] reported a shorter mean interval to consultation of 6 months. Thus, in case of chronic otorrhea resistant to treatment for infection, and especially when otoscopy finds a polyp or external auditory canal collapse, CT assessment should be undertaken without delay [5]. In the present case, osteolysis seen on CT imaging suggested middle-ear cholesteatoma. A diagnosis of middle-ear angiosarcoma presupposes considerable experience on the part of both the clinician and the radiologist. Only anatomopathological examination of a biopsy sample taken from the suspect lesion enables diagnosis to be confirmed. In the present case, diagnosis was confirmed only by histologic examination of the surgical specimen. Table 1 presents the cases of middle-ear angiosarcoma retrieved from the literature [7,8]. Although there is no consensus, the 1990 Pittsburgh TNM classification of external auditory canal squamous cell carcinoma [9,10] is the most widely used. It is based on preoperative clinical examination and CT findings [9], and has the advantage of being of both therapeutic and prognostic interest. A minor revision of the classification was made in 2000 [9].

Treatment of middle-ear tumor depends on the degree of extension [3]. Classically, it associates surgery and/or radiochemotherapy. Surgery should follow the rules of cancer surgery rather than the general principles of ear surgery [5]. It should be large-scale and adapted, generally consisting in petrosectomy and requiring a dedicated technical platform [6,10]. In the present case, large-scale petromastoid dissection allowed satisfactory tumor removal. The prescribed radiochemotherapy could not be carried out, radiotherapy not being available in Côte d'Ivoire. Only adjuvant chemotherapy could be administered, following the usual protocol. Prognosis depends on the interval to diagnosis and on tumor extension. In evolved forms, surgical removal is incomplete in 50% of cases, accounting for the high rate of local recurrence at 10 months [3]. In the present case, there was no local recurrence at 12 months' follow-up.

Conclusion

Middle-ear angiosarcoma is a rare tumor, like other middle-ear tumors. Diagnosis is delayed by the non-specificity of early signs, leading the otologist to suspect an infectious etiology. CT is of great diagnostic interest, on condition that the radiologist be sufficiently experienced. Histologic examination of a biopsy sample, to be taken from any suspect lesion, is the key to diagnosis and the only guarantee of effective treatment. We would stress the need for multidisciplinary management associating otologist, radiologist and oncologist.

Conflict of interest statement

None.

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